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【 CASE REPORT 】

Can Takayasu Arteritis Cause Hydronephrosis?

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Abstract:

A 74-year-old woman was admitted because of malaise and a low-grade fever. Her C-reactive protein level was 0.96 mg/dL. Computed tomography (CT) revealed diffuse uniform thickening of the arterial wall from the abdominal aorta to the common iliac artery and right hydronephrosis. ^{18}F-fluordesoxyglucose positron emission tomography-CT showed an accumulation in the same area. These findings suggested Takayasu arteritis and retroperitoneal fibrosis as differential diagnoses. Takayasu arteritis is characterized by thickening of the arterial walls, and retroperitoneal fibrosis is characterized by membranous lesions covering the outer surface of the arterial walls. Thus, Takayasu arteritis was deemed the most likely diagnosis. Steroid treatment was effective.

Key words: Takayasu arteritis, retroperitoneal fibrosis, hydronephrosis, ^{18}F-fluordesoxyglucose positron emission tomography

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Introduction

Takayasu arteritis (TA) is a large-vessel vasculitis that affects the aorta and its primary branches, coronary arteries, and pulmonary arteries. Its principal manifestations are systemic inflammation, which may be characterized by a fever and elevated C-reactive protein (CRP) level, and pain, which may be caused by arterial inflammation; symptoms related to vascular narrowing, occlusion, and dilation are also seen. Histologically, the initial lesion begins with inflammation of the outer membrane of aorta, which causes edematous changes and thickening of the outer membrane. The inflammation then affects the tunica media, where it damages the elastic fibers, followed by the inner membrane, eventually leading to thickening of the inner membrane and luminal narrowing (1-3). TA is most common in young women. An early diagnosis is known to be challenging, but regions affected by TA can now be detected by diagnostic imaging, including contrast-enhanced computed tomography (CT) and contrast-enhanced magnetic resonance imaging (MRI), which show thickening of the aortic wall, as well as ^{18}F-fluordesoxyglucose positron emission tomography (FDG-PET)-CT, which shows the accumulation of ^{18}F-FDP in affected lesions (1).

TA may affect the kidneys in the form of renal artery stenosis due to spreading of aortic inflammation. However, to our knowledge no report has described a case of TA concomitant with hydronephrosis due to inflammatory thickening of a common iliac lesion that affects the adjacent ureter.

We herein report a hydronephrosis patient with TA and discuss how the two conditions are connected.

Case Report

A 74-year-old Japanese woman was admitted to our hospital for the evaluation of right hydronephrosis. The patient had no history of hospital visits because she had had no previous subjective symptoms. One year before admission, she became aware of malaise, a low-grade fever, and pain in both upper limbs and bilateral proximal thighs, and three months before admission, she visited a primary doctor for

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gastric discomfort. At that time, right hydronephrosis was detected. She had no history of hypertension or smoking.

On admission, the patient was 153 cm tall and weighed 48.2 kg. Her blood pressure was 147/78 mmHg in the right arm and 145/75 mmHg in the left arm, and her heart rate was 75 beats/min with a body temperature of 37.1°C. She complained of bilateral femur pain, but no edema of the lower extremities, retinopathy, or peripheral neuropathy was noted. There was no evidence of enlargement of the submandibular, parotid, or lacrimal glands.

Laboratory findings were as follows: white blood cell count, 6,000/μL; red blood cell count, 4.28×10^{6}/μL; hemoglobin, 11.3 g/dL; platelet count, 360×10^{3}/μL; total protein, 7.2 g/dL; albumin, 3.7 g/dL; serum urea nitrogen, 22 mg/dL; serum creatinine, 1.16 mg/dL; estimated glomerular filtration rate (eGFR), 35.4 mL/min/1.73 m^2; CRP, 0.96 mg/dL; erythrocyte sedimentation rate (ESR9, 84 mm/h; IgG, 1,402 mg/dL; IgG4, 35 mg/dL; IgA, 348 mg/dL; IgM, 110 mg/dL); antinuclear antibody, negative; myeloperoxidase antineutrophil cytoplasmic antibodies, less than 0.5 IU/mL; and proteinase 3 antineutrophil cytoplasmic antibodies, <0.5 IU/mL. Total urinary protein excretion was 0.2 g/day, and the sediment contained 1 to 4 erythrocytes per high-power field.

Ultrasonography showed right common iliac arterial wall thickening of 3.3 mm and right hydronephrosis. CT revealed diffuse uniform thickening of the arterial wall from the abdominal aorta to the common iliac artery, which was clearly visible after administration of contrast medium, as well as dilation of the ureter from the renal pelvis to the vicinity of the common iliac artery (Fig. 1a-d). No pancreatic enlargement was observed. FDG-PET-CT scan showed the accumulation in the thickened region of the aortic wall (Fig. 1e-g). No stenosis of the internal carotid artery, brachial artery, or renal artery was observed. No lesions showing significant uptake were found in other organs.

The diagnosis

TA was diagnosed based on the evidence of an inflammatory reaction in the blood tests and diagnostic imaging findings. After the ureteral stricture was located by insertion of a ureteral stent, the site of the stricture and the CT and PET-CT findings were reviewed by a urologist and a radiologist. Anatomically, the right and left ureters cross over the ventral side of the right and left common iliac arteries, respectively. In this case, we speculated that the right iliac artery was thickened because of vascular inflammation, entrapping the right ureter and resulting in ureteral stricture and hydronephrosis.

Clinical course

Treatment with 30 mg/day prednisolone was started 4 days after admission (day 4). On day 11, CRP was 0.09 mg/L, ESR was 22 mm, and the patient's bilateral femoral pain had resolved. On day 14, ultrasonography revealed that the common iliac artery wall width had improved from 3.3 mm to 1.0 mm, and mild improvement of hydronephrosis was also noted. On day 130, the ureteral stent was removed. On day 268, CT images showed no hydronephrosis and a normal aortic wall (Fig. 2).

Discussion

We encountered a case of TA with hydronephrosis. Previously, the most common renal manifestations of TA were described as renovascular hypertension and ischemic nephropathy due to stenosis and occlusion of the renal artery and its branches (4). Only a few clinical reports have described glomerular changes associated with TA, including diffuse mesangial proliferative glomerulonephritis (GN), focal segmental glomerulosclerosis, global and focal glomerulosclerosis, segmental necrotizing GN, amyloidosis, and membranous nephropathy (5-8). However, hydronephrosis has been reported rare as a clinical symptom of TA.

To reach a diagnosis, we reviewed the differential diagnoses of aortic lesions that cause hydronephrosis. Retroperitoneal fibrosis (RPF) is the most common disease that should be differentiated. It is characterized by the presence of fibroinflammatory tissue, which usually surrounds the abdominal aorta and iliac arteries and extends into the retroperitoneum to envelop neighboring structures, including ureters, resulting in hydronephrosis (9). However, RPF does not directly affect the aortic wall. The disease is generally divided into two types: idiopathic RPF without identified pathogenesis, and secondary RPF. Recent studies on IgG4-related disease revealed that biopsies of affected organs show abundant infiltration of IgG4-positive plasma cells. A biopsy of the retroperitoneal tissue is rarely performed, and the diagnosis is made by a biopsy of lesions in other organs, such as salivary glands and kidneys (10). In our case, no IgG4-specific organ lesions, such as major salivary gland enlargement, renal lesions, or pancreatic lesions, were identified on PET-CT or other imaging modalities.

Baldwin et al. studied 122 patients who showed complications of renal artery involvement due to TA. They reported that in two patients, aortic lesions caused stenosis in the transition zone of the renal pelvis and ureter, resulting in hydronephrosis. This suggests that abdominal aortic lesions may entrap the ureter in the common iliac region and cause hydronephrosis (11).

There are reports that inflammatory lesions of retroperitoneal fibrosis are also present in the aortic wall. Adnan et al. performed PET on 12 patients with retroperitoneal fibrosis, where the ^{18}F-fluorodeoxyglucose-avid uptake suggested inflammatory lesions. Thirteen patients had positive findings in the periarterial soft tissue, and in five cases, partial or total positive findings were also found in the arterial wall. This suggests that inflammation in RPF occurs not only in the soft tissue around the arteries, but also in the arteries themselves. In contrast, the positive findings on PET were localized only at the arterial wall, which is characteristic of TA. While the two diseases involve the aorta, the main dif-

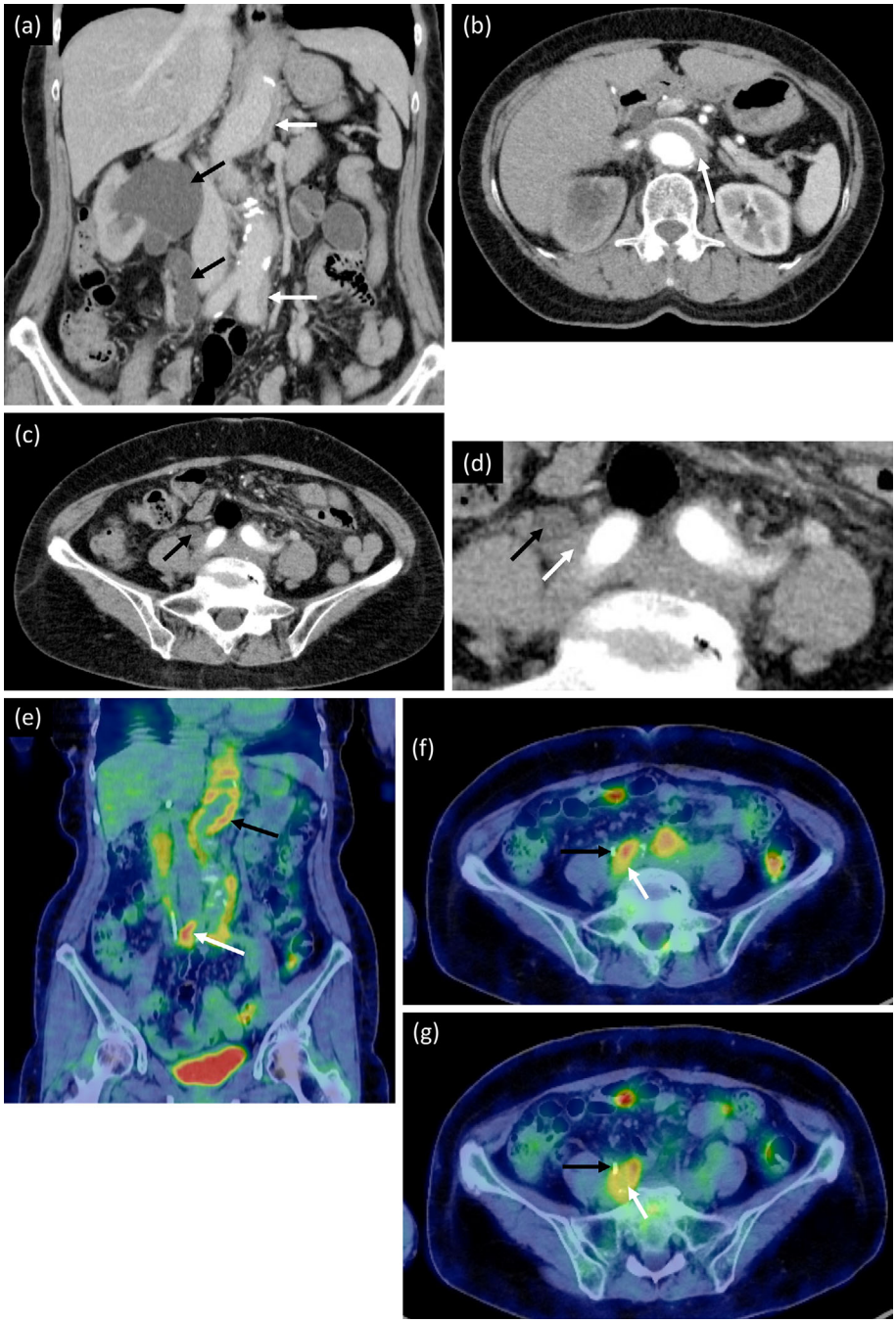

Figure 1. **Initial contrast-enhanced computed tomography scan after administration of contrast medium. a: The image shows diffuse uniform thickening (white arrowhead) of the arterial wall from the abdominal aorta to the common iliac artery and dilation of the renal pelvis and ureter (black arrowhead). b: Diffuse uniform thickening (white arrowhead) can be seen in the abdominal aortic wall. c: Dilation of the ureter (black arrowhead) continues to the vicinity of the common iliac artery. d: Magnified image of (c). The black arrowhead shows dilation of the ureter. The white arrow shows the thickened common iliac artery wall. e: ^{18}F-fluordesoxyglucose positron emission tomography/computed tomography shows the accumulation in the thickened region of the aortic wall (black arrowhead) and the common iliac artery (white arrowhead). f: ^{18}F-fluordesoxyglucose positron emission tomography/computed tomography shows the accumulation (white arrowhead) in the thickened region (yellow color) of the common iliac arterial wall. The black arrowhead shows the ureteral stent (white color) just before entering the arterial wall. g: ^{18}F-fluordesoxyglucose positron emission tomography/computed tomography shows the accumulation (white arrowhead) in the thickened region (yellow color) of the common iliac arterial wall. The black arrowhead shows the ureteral stent (white color) entering the arterial wall.**

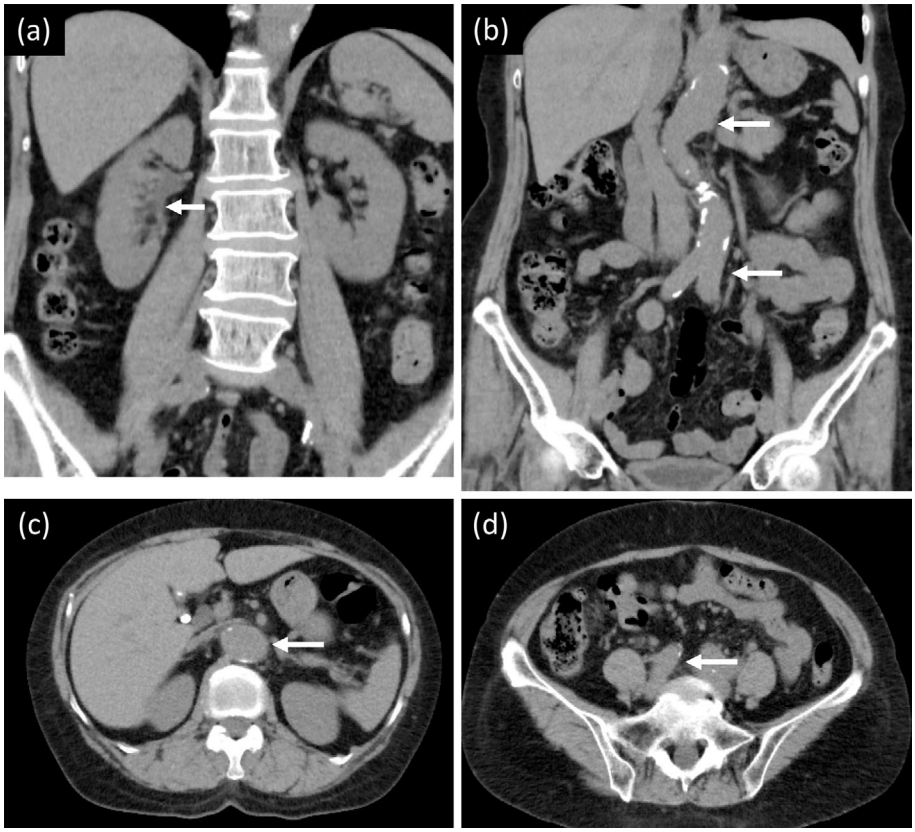

Figure 2. Computed tomography findings after treatment. a: Right hydronephrosis improved (white arrowhead). b: Wall thickening from the abdominal aorta to the common iliac artery improved (white arrowhead). c: Wall thickening of the abdominal aorta improved (white arrowhead). d: Wall thickening of the common iliac artery improved (white arrowhead).

ferentiating feature may be whether or not the inflammation extends outside the artery as well (12).

The main differential diagnosis for "disease with the abdominal aorta as the site of inflammation" is inflammatory abdominal aortic aneurysm (IAAA), which is characterized by aneurysms and markedly thickened arterial walls that are enhanced after contrast media use, and fibrosis of the adjacent retroperitoneum and rigid attachment of adjacent structures to the anterior aneurysmal wall are additional features. The lack of significant aneurysm with TA suggests that differentiation is possible (13). Differentiating between the three diseases (TA, RPF, and IAAA) is considered important for the etiology of TA.

We considered TA to be the most appropriate diagnosis for this case, excluding RPF and IAAA, but the patient's older age of onset differed from the typical TA course. The development of hydronephrosis, which is not usually seen in young-onset TA, may have been related to factors associated with older-onset TA.

In summary, we experienced a case in which inflammatory aortitis was the lesion responsible for hydronephrosis. The most likely diagnosis in a case of an aortic lesion presenting with hydronephrosis is RPF. However, the imaging studies showed findings typical of TA rather than RPF. Although TA complicated by hydronephrosis has not previously been reported, anatomical considerations suggest that outward wall thickening of the common iliac artery due to inflammation of the adventitia may result in hydronephrosis by causing ureteral stricture.

This case study was performed in accordance with the Declaration of Helsinki. The patient provided her written informed consent for the publication.

The authors state that they have no Conflict of Interest (COI).

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